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Case report

Cardiac tamponade secondary to fulminant myocarditis - A case of custodial death

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ABSTRACT

Cardiac tamponade following rupture of the heart occurs very rapidly, resulting in a fatal fall in the cardiac output and circulatory collapse. Spontaneous cardiac rupture is an uncommon occurrence and that too occurring secondary to myocarditis is a very rare event. Myocarditis is an inflammatory disease of the myocardium and its clinical presentation is highly variable. Due to its highly variable clinical presentation, the diagnosis is frequently made at autopsy. In this article, we report death of a prisoner due to cardiac tamponade following right ventricular rupture secondary to fulminant myocarditis.

1. Introduction

Myocarditis is an inflammatory disease of the myocardium with a wide range of clinical presentation. Due to its highly variable clinical presentation, the diagnosis is frequently made at autopsy. Myocarditis is defined as "a process characterized by an inflammatory infiltrate of the myocardium with necrosis and/or degeneration of adjacent myocytes not typical of ischemic damage associated with coronary artery disease.[1] Autopsy studies report a frequency of myocarditis ranging from 0.11 to 5.55% in the general population.[2] Spontaneous cardiac rupture leading to cardiac tamponade is one of the fatal outcomes of fulminant myocarditis. In this paper we report a fatal case of cardiac tamponade secondary to rupture of heart in a prisoner suffering from fulminant myocarditis. In cases of custodial deaths, meticulous autopsy not only helps in the determination of actual cause and manner of death but also assists in putting an end to the suspicions surrounding the death of the deceased.

2. Case history

A 52 years old man was brought to the casualty with complaints of sudden onset of chest pain followed by loss of consciousness since 1 h. On examination, engorged jugular veins, very distant heart sound, coarse breathing sound bilaterally, cyanotic extremities, reactive pupil, feeble peripheral pulse and systolic blood pressure 60 mm Hg. Immediately he was put on ventilator. Unfortunately, the patient expired before further investigations were made. According to the law of the land, body was subjected for medico legal autopsy.

Patient had been apparently healthy except for flu like symptoms one week prior to admission for two days. He was a chronic smoker; but, non alcoholic, non hypertensive and non diabetic. There was no clinical history of previous ischemic heart disease. There was no family history of cardiac disorder.

3. Autopsy findings

On external examination, there was no significant finding apart from the postmortem changes corresponding to the time of death; and no evidence of infliction of violence. On internal examination, positive findings were confined to the cardiovascular

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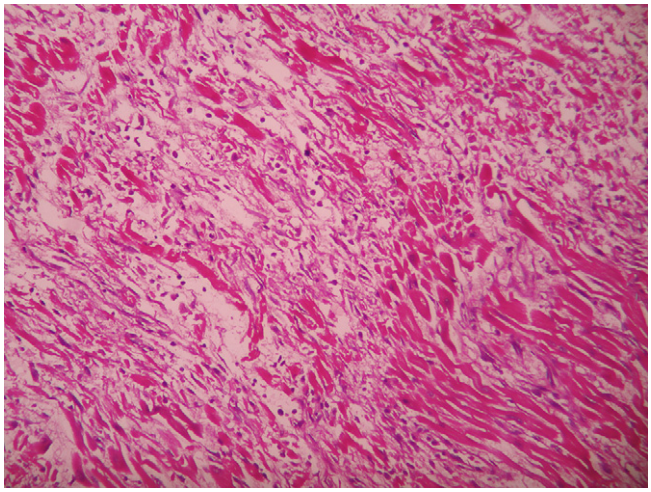

Fig. 1. Microphotograph from the right ventricle, showing Myocardial fiber disarray and Chronic inflammatory infiltrate (H & E, × 200).

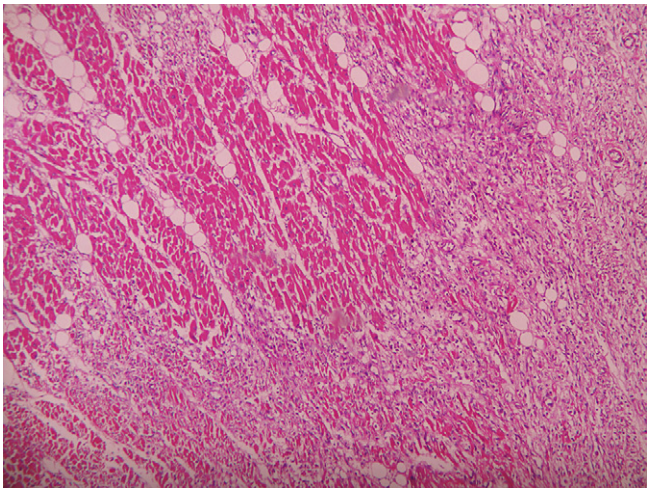

Fig. 3. Microphotograph from the inter-ventricular septum, showing Myocardial fiber disarray and Chronic inflammatory infiltrate (H & E, × 100).

system. There was 400 ml of fluid blood in the pericardial sac. A rupture of size 3 mm in diameter with hemorrhagic frayed edges was present in the right ventricle anteriorly. Blood clot weighing 27 gm was present over the rupture. There was no evidence of pale or necrotic areas suggestive of myocardial infarction. Cardiac valves and chambers were normal. The coronary arteries showed only minimal bright yellow atheromatous plaques without any occlusion. There was no cardiomegaly and cardiac hypertrophy. There were no congenital and acquired anomalies of the heart. Heart weighed 339 gm. Other viscera were intact and did not show any injury or anomaly. On cut section all organs were congested. All organs were sent for histo-pathological examination.

On microscopy, rupture was present in the right ventricle anteriorly. Myocardium of the right ventricle (Figs. 1 and 2), inter-ventricular septum (Figs. 3 and 4), left ventricular wall around the coronary sinus (Figs. 5 and 6) showed focal areas of myocarditis showing myocardial fibers disarray with myocytolysis. These areas were extensively infiltrated by lymphocytes. Tissues from the left and right atrial wall revealed normal myocardium. There was no evidence of myocardial infarction. Pericardium also showed infiltration of lymphocytes and plasma cells suggestive of pericarditis (Fig. 7). Coronaries showed Grade II atherosclerosis without significant narrowing of the lumen. Pathological diagnosis was fulminant myocarditis with rupture in right ventricular wall. All the other organs showed congestion.

On perusal of clinical history, autopsy findings and histo-pathological examination report, cause of death was opined as "Cardiac tamponade following spontaneous rupture of right ventricle secondary to fulminant myocarditis" and manner of death as "Natural".

4. Discussion

Spontaneous cardiac rupture is a rare occurrence. Most of the cases reported in the literature were caused by myocardial infarction. Some sporadic cases happened in myocarditis, mediastinitis, myocardial abscess, angiosarcoma of the heart, Chaga's disease and after mitral valve replacement surgery. Cardiac tamponade following rupture of the heart occurs very rapidly, resulting in a fatal fall in the cardiac output and circulatory collapse. The interval between rupture and collapse may be variable, but it is usually short, even when the rupture hole is small.[3]

Rupture of myocardium is classified into three types: 1. Blow up rupture in which the patient usually dies within minutes before

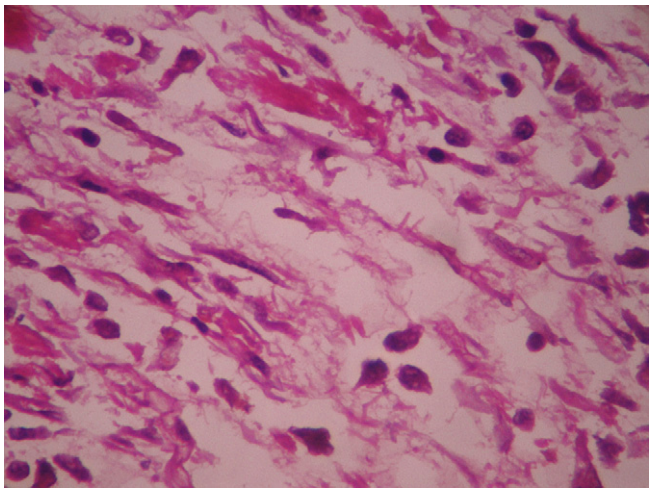

Fig. 2. Microphotograph from the right ventricle, showing Myocytolysis and Inflammatory infiltrate with lymphocytes and plasma cells. (H & E, × 400).

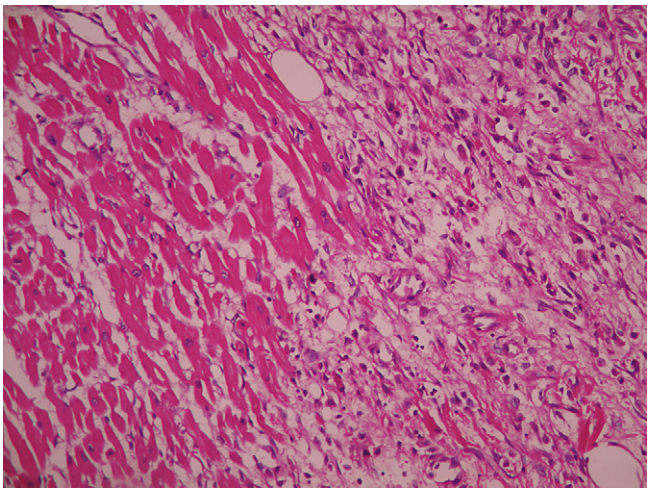

Fig. 4. Microphotograph from the inter-ventricular septum, showing Myocardial fiber disarray, myocytolysis and Chronic inflammatory infiltrate (H & E, × 200).

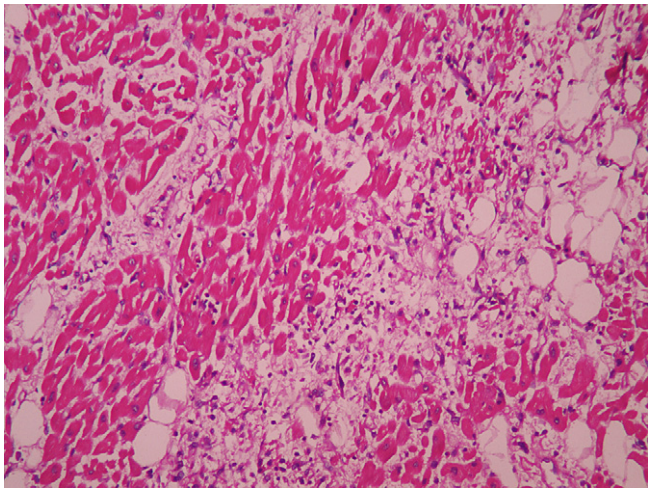

Fig. 5. Microphotograph from the left ventricular wall (1 cm below the A-V Junction) showing Myocardial fiber disarray and Chronic inflammatory infiltrate (H & E, × 200).

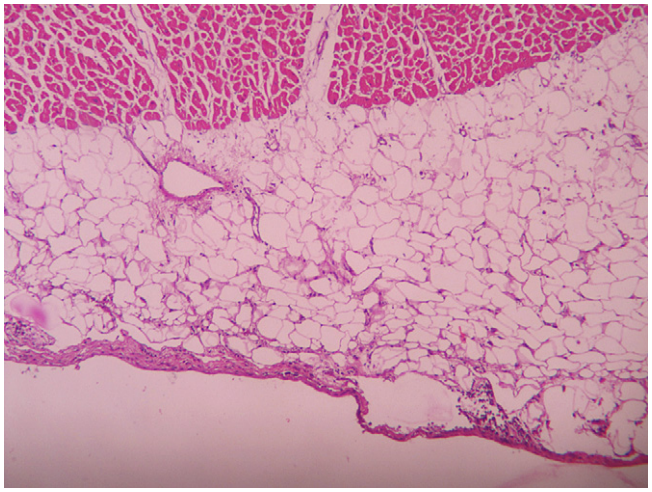

Fig. 7. Microphotograph from the pericardium, showing pericarditis with few myocardial fibers, epiardial adipose tissue infiltrated with chronic inflammatory cells and edema at one focus. (H & E, × 200).

arriving at the hospital; 2. Small rupture which makes an operation possible if proper intervention is carried out within a few hours with the help of adequate hemodynamic support; and 3. Chronic rupture, which will have formation of false aneurysm.[3]

Myocarditis is defined as "a process characterized by an inflammatory infiltrate of the myocardium with necrosis and/or degeneration of adjacent myocytes not typical of ischemic damage associated with coronary artery disease". Necrosis of the myocardium can lead to rupture. Myocarditis requires an inflammatory infiltrate and damage to the adjacent myocytes confirmed by light microscopy. Myocarditis is classified into primary and secondary myocarditis. Primary myocarditis is presumed to be due to either an acute viral infection or a post viral autoimmune response. Secondary myocarditis is myocardial inflammation caused by specific pathogens like bacteria, spirochetes, rickettsia, fungi, protozoa, drugs, chemicals, physical agents and other inflammatory diseases like systemic lupus erythematosus. Primary myocarditis is further classified into fulminant, acute (sub acute), chronic active and chronic persistent myocarditis. The present case has distinct onset of the condition following an episode of viral fever, multiple foci of inflammatory infiltrates and myocyte necrosis; and it fits into the class of fulminant Myocarditis.[1]

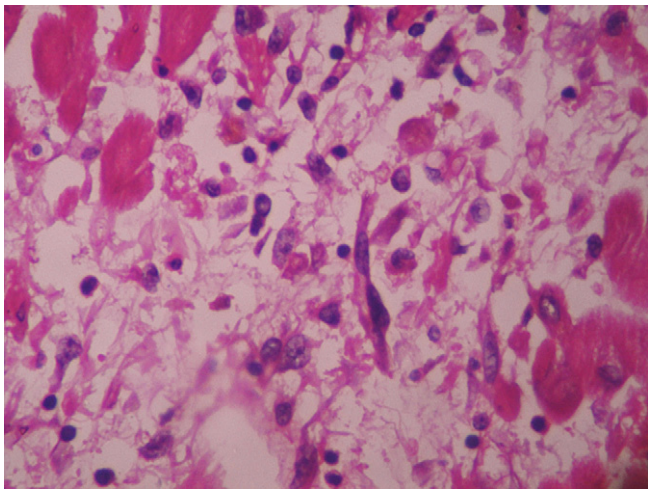

Fig. 6. Microphotograph from the left ventricular wall (1 cm below the A-V Junction) showing Myocytolysis and Inflammatory infiltrate with lymphocytes and plasma cells. (H & E, × 400).

Myocarditis is classified as idiopathic in approximately 50% of the cases. Klugman et al found that 82% of the pediatric cases studied were considered idiopathic.[4] The investigators also determined that 3% of cases in the study had a known bacterial or viral etiology, and that 6% of cases were related to other diseases. In idiopathic cases, a viral etiology is often suspected but unproved, even with sophisticated immunohistochemical and genomic studies.[5]

In myocarditis the myocardial damage has two main phases: 1. Acute Phase (first 2 weeks): In this phase, the myocyte destruction is a direct consequence of the offending agent, which causes cell mediated cytotoxicity and cytokine release, contributing to myocardial damage and dysfunction. Detection of causal agent is uncommon during this stage; and 2. Chronic Phase (more than 2 weeks): Continuing myocyte destruction is autoimmune in nature, with associated abnormal expression of human leukocyte antigen in myocyte (and in the case of viral myocarditis, persistence of viral genome in myocardium).[6]

Clinical presentation of myocarditis is variable, ranging from an asymptomatic or self limited disease to profound cardiogenic shock. Majority of the patients have no specific cardiovascular abnormalities, but may have ST segment and T wave abnormalities on electrocardiogram. Chest pain may occur in up to 35% of patients and this usually reflects associated pericarditis.[7]

Endomyocardial biopsy is an important diagnostic tool in the clinically suspicious myocarditis cases. In 1984, the Myocarditis panel met in Dallas with the major purpose of developing practical set of histo-pathological criteria for the diagnosis of myocarditis on endomyocardial biopsies. The criteria laid down by this panel are known as Dallas Criteria. Dallas criteria myocarditis requires an inflammatory infiltrate and associated myocyte necrosis or damage not characteristic of an ischemic event. Borderline myocarditis requires a less intense inflammatory infiltrate and no light microscopic evidence of myocyte destruction.[8] These criteria have been used in the histo-pathological diagnosis of clinically suspected cases of myocarditis over the last two decades. However, the recent studies suggest that the Dallas criteria are no longer adequate in the diagnosis of myocarditis due to sampling error, variation in expert interpretation, variance with other markers of viral infection and immune activation in the heart and variance with treatment outcomes.

Chow et al and Hauck et al demonstrated by biopsying post-mortem hearts of patients who had died with myocarditis that, from a single endomyocardial biopsy, histological myocarditis could be demonstrated in only 25% of samples. With more than 5 biopsies, Dallas criteria myocarditis could be diagnosed in approximately two thirds of subjects. The authors, of a recent MRI study used focal imaging abnormalities to guide heart biopsy investigation of possible myocarditis, showed that the earliest myocardial inflammatory abnormalities were evident in the lateral wall of the left ventricle, and only these sites revealed myocarditis by histological examination.[9] Hence, there is significant sampling error associated with establishing the diagnosis of myocarditis. Also, there are variations in the interpretation of histo-pathological samples. In a multi-institutional Myocarditis Treatment Trial,[10] of the 2000 plus subjects entered in the trial with clinical suspicion of myocarditis, only 111 patients had myocarditis based on the Dallas criteria as applied by the local pathologists. Furthermore, of the 111 patients only 66 were diagnosed as having Dallas criteria myocarditis by the expert pathology panel who reviewed the histo-pathological material. In another separate analysis,[11] seven expert pathologists' interpretations of histo-pathological findings from endomyocardial biopsies of 16 patients with dilated cardiomyopathy varied remarkably in the diagnosis of myocarditis. Definite or probable myocarditis was diagnosed in 11 of 16 patients by at least 1 pathologist. However, of the 11 patients, 3 of 7 pathologists agreed on the diagnosis of myocarditis in 3 patients, and 2 of 7 pathologists agreed on the diagnosis of myocarditis in 5 patients. Therefore, even expert observers are not of the same opinion in the interpretation of histo-pathological material. Patients with "primary" (or post viral) myocarditis constitute the larger and more important category of myocarditis. Many studies have shown that virus may be present in the myocardium without Dallas criteria myocarditis. Virus can exist in the myocardium (even in a replicative form) in the absence of myocardial inflammation adequate to meet Dallas criteria and may adversely affect outcome. Martin et al[12] demonstrated in 34 children with clinical presentations compatible with myocarditis that 26 heart biopsy samples were positive for viral pathogens, and 13 of the 26 positive samples had no evidence of myocarditis by histo-pathological examination. A meta-analysis[9] of polymerase chain reaction studies in patients who had heart biopsies with presumed myocarditis or cardiomyopathy demonstrated an odds ratio of 3.8 for viral presence in both categories compared with control patients. Therefore, the limitations of the Dallas criteria like variability in interpretation, sampling error etc and the fact that the clinically suspicious myocarditis is a heterogenous disorder caused by a myriad of mechanisms, have led to the application of Polymerase Chain Reaction technique to determine virus involvement, immunohistochemical stains of endomyocardial biopsies for leucocytes and surface antigens, such as Inter Cellular Adhesion Molecule (ICAM) or Human Leucocyte Antigen - DR as well as serum immunological studies for circulating antimicrobial antibodies. It is the need of hour for the clinicians, pathologists, immunologists and molecular cardiologists to contribute to the new criteria, which should include clinical presentation, histopathology, immunohistochemistry, viral polymerase chain reaction, cardiac antibody assessment and imaging results.

Epidemiology of fatal myocarditis in general population has remained largely un-characterized, because the clinical presentations and findings of myocarditis are highly variable and the definite diagnosis is based on autopsy. Autopsy studies report a frequency of myocarditis ranging from 0.11 to 5.55% in the general population.[2] In a retrospective analysis of autopsy cases entered in the Annuals of Autopsy Records for Japan from 1958 to 1977, 434 cases of myocarditis (0.12%) were present out of total 3,77,841 cases.[13] In another study conducted at Finland, myocarditis was recorded to cause 0.47 out of 1000 deaths in the general Finnish population during the years 1970–1998.[14]

Due to the rising incidents of deaths in police lock up and jails, The National Human Rights Commission of India has laid down stringent guidelines for the investigation of such deaths. In the present case, the deceased was a prisoner and hence, the determination of manner of death was equally important as finding out the cause of death; because, further investigation largely depends upon the manner of death.

5. Conclusion

Cardiac tamponade following rupture of heart in an individual suffering from fulminant myocarditis is a rare event. Myocarditis has highly variable clinical presentation and hence, the diagnosis is frequently made at autopsy. The present case once again stresses the importance of meticulous autopsy with all possible investigations in cases where cause and manner of death is in question and more so in custodial deaths. In recent times due to the increase in the number of custodial deaths, the common dictum is to consider all such deaths as unnatural until the contrary is proved.

Conflict of interest statement

The authors declare that there is No Conflict of Interest.

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